Life Expectancy Without Surgery in Tetralogy of Fallot

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All published autopsy cases of patients with tetralogy of Fallot who died without surgical treatment were studied to determine the life expectancy of such persons. In addition, the data from a study of persons with tetralogy alive in Denmark in 1949 were reanalyzed. The survival data from these two sources were remarkably similar, indicating that 66 percent of persons with tetralogy of Fallot not treated surgically live to age 1 year, 49 percent to age 3 years and 24 percent to age 10 years; thereafter, the hazard function (or instantaneous risk of death) remains constant. The chance of survival is significantly less when pulmonary atresia, rather than stenosis, is present.

Surgical treatment is indicated for most patients with tetralogy of Fallot and is usually effective. However, several questions remain unanswered, including how many infants with tetralogy should be treated surgically and whether patients with tetralogy who survive into the third and fourth decades of life require an operation. Data on life expectancy and other aspects of the natural history of persons with tetralogy of Fallot are required to answer these questions.

Randomized studies of the length of life with and without surgical treatment in patients with tetralogy of Fallot are not now possible. Therefore, we analyzed published clinical and autopsy data to estimate the life expectancy of persons with tetralogy who were not treated surgically.

Material and Methods

The 1949 Danish study: The material for our study came from two sources. The first was from Rygg et al.,[1] who determined the ages of all persons with tetralogy of Fallot alive in Denmark in 1949. That year was chosen because until 1949 no person in Denmark had had corrective surgery for tetralogy of Fallot. Fifty-four of the 216 persons with tetralogy had had palliative operations in 1948 and 1949, and 5 had died at operation. Most of these 54 patients were older than age 6 years and all were older than age 2 years at operation. For the purposes of the study, all 54 patients were considered to be alive at the end of 1949. The number of births in Denmark in each of the 58 previous years was known. It was assumed in the study that each year tetralogy of Fallot was present in 0.4 of every 1,000 live births. From these data the number of persons born with tetralogy of Fallot who were alive in 1949 at ages 1 to 59 years was calculated and their length of life estimated.

Published autopsy cases: Our second source of data was reported autopsy cases of tetralogy of Fallot. Most cases included were reviewed individually in their original publication to determine whether the diagnosis was in fact tetralogy of Fallot, whether the age at death was known and, as far as possible, whether the death was related to the presence of the malformation. However, some data from reliable sources were presented as "grouped" information (for example, "10 cases dying between ages 1 to 10 years"), and thus the individual cases could not be examined. In 54 cases sufficient data were presented to indicate that the

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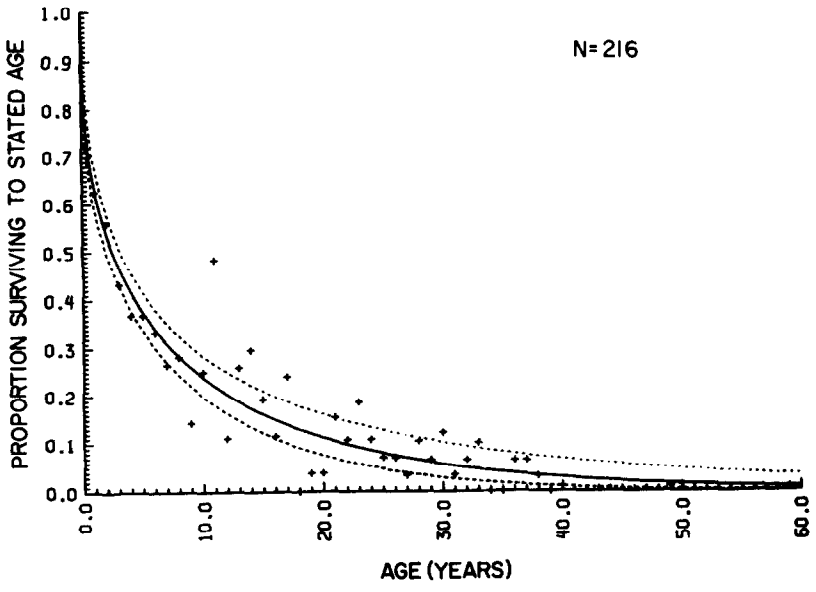


FIGURE 1. Life expectancy of persons with tetralogy of Fallot, based on data from the Danish population study. **Crosses** represent data points calculated in that study. The **solid line** (with its 70 percent confidence limits enclosed by the **dashed lines**) represents parametric analysis of the data, which resulted in the equation (single hit model[19]): Probability of dying = $[1 - \exp(\mu t/m)]^m$, where exp is e, the base of the natural logarithms; $\mu = \exp(\beta)$ where $\beta = -6.2 \pm 0.25$; t is age on January 1, 1950 in months and $m = 0.36 \pm 0.056$. Significance levels of coefficients, $P_\beta < 0.0001$ and $P_m < 0.0001$; correlation between coefficients, $r = 0.976$. The results are shown to age 60 years. N = number of patients.

diagnosis was tetralogy of Fallot with pulmonary atresia.[2–8] In 195 cases sufficient data were found to indicate that the diagnosis was tetralogy of Fallot without pulmonary atresia.[2–4,6,9–13] These cases, in addition to 317 cases in which the presence or absence of pulmonary atresia could not be determined, constitute the total group of 566 autopsy cases.[2–17]* The autopsy cases were analyzed to determine survival or life expectancy data, that is, the cumulative number of cases in which the age at death was greater than a series of ages, t.

Survival curves of cases with pulmonary stenosis versus atresia: The survival data in cases without pulmonary atresia appeared more favorable in the early years of life than current clinical experience suggests. Most of the 195 cases in this group were reported by persons and institutions known to care for few if any small children. Furthermore, when the 195 cases without and the 54 cases with pulmonary atresia are removed from the total group of 566 cases, the survival data of the remaining cases are much less favorable than the data from the total group. This suggests that the 195 patients removed were a particularly favored subset among those without pulmonary atresia. Also, if these data are correct, mathematical analysis of the combined curve indicates that 60 percent of the patients at any age had pulmonary atresia, which is highly unlikely. Because these data appeared unreliable, we looked for another method of determining the life expectancy of persons with tetralogy of Fallot but without pulmonary atresia. Knowing the survival data for persons with pulmonary atresia, assuming a 33 percent incidence rate of

* The specific case numbers in each publication and the original source for each case in review series are available from the authors.

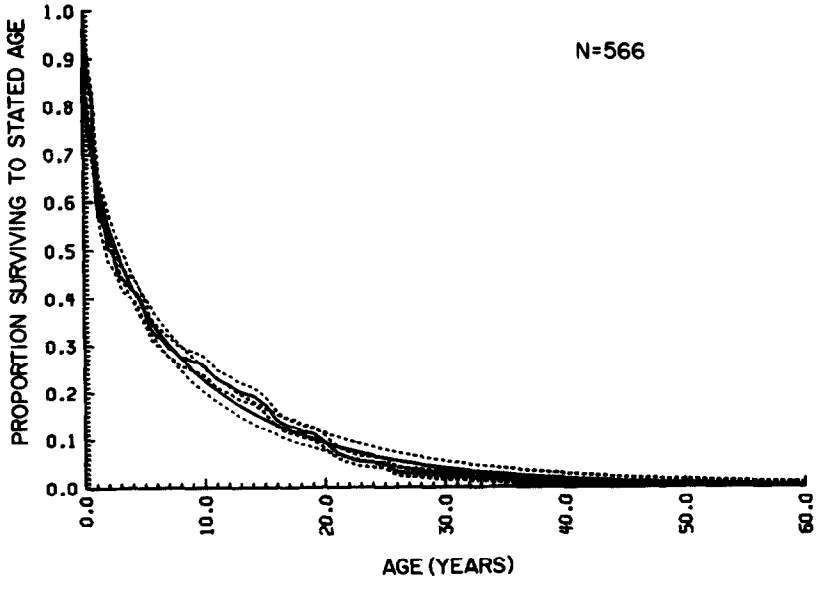
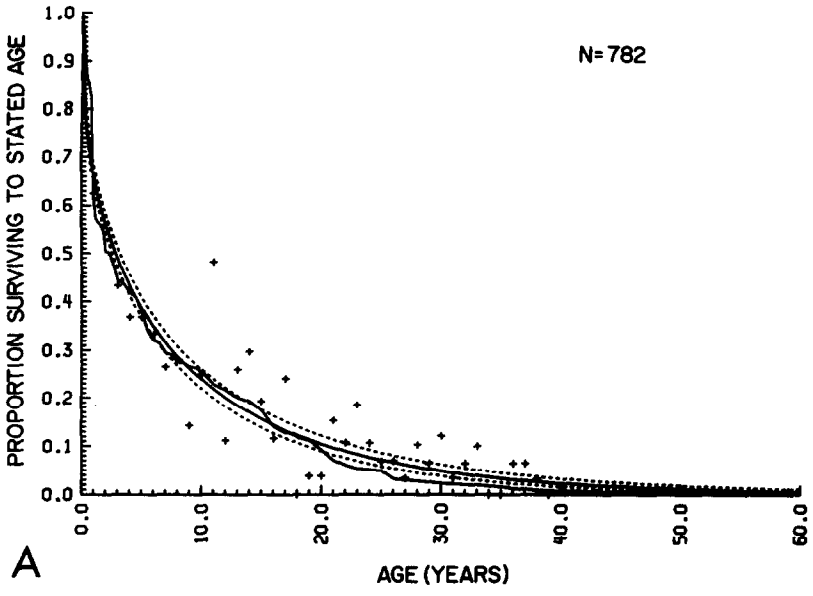
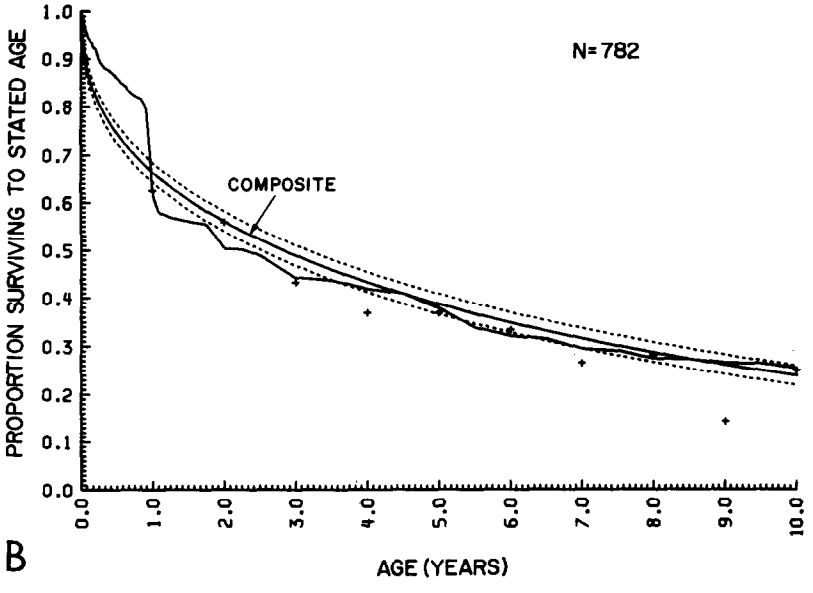


FIGURE 2. Life expectancy of persons with tetralogy of Fallot, based on autopsy study.[2–17] The **jagged line** (with its 70 percent confidence limits enclosed by the **dashed lines**) represents the data from the nonparametric actuarial analysis. The **smooth line** (with its 70 percent confidence limits) represents the parametric analysis, which yielded the same equation as in Figure 1, with the coefficients $\beta = -5.87 \pm 0.081$ and $m = 0.425 \pm 0.0178$. Significance level of the coefficients, $P_\beta < 0.0001$ and $P_m < 0.0001$; correlation between coefficients: $r = 0.500$. N = number of patients.





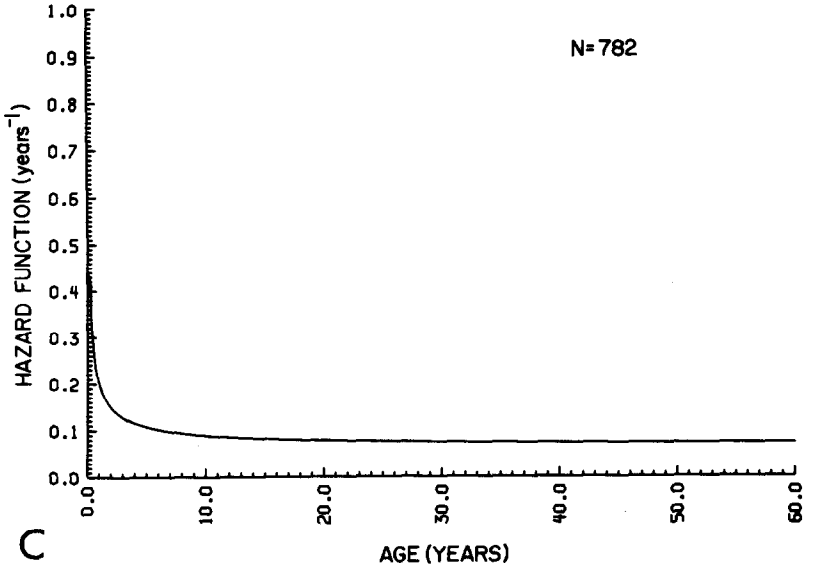


FIGURE 3. Life expectancy of persons with tetralogy of Fallot, based on combined analysis of the Danish data and autopsy data. **A,** results to age 60 years; **B,** results to age 10 years on an expanded time scale; **C,** proportion of persons at risk of dying each year (hazard function), according to the age of the person. The greatest risk is in the first year of life. The actual data points (**crosses**) calculated in the Danish study are shown, as is the nonparametric actuarial analysis (**jagged line**) from the autopsy data. The smooth line (with its 70 percent confidence limits enclosed by the **dashed lines**) results from a combined parametric analysis that yielded the same equation as in Figure 1 with the coefficients, $\beta = -6.00 \pm 0.072$ and $m = 0.410 \pm 0.0172$. Significance level of the coefficients, $P_\beta < 0.0001$ and $P_m < 0.0001$; correlation between coefficients, $r = 0.809$.

congenital pulmonary atresia among 566 autopsy cases of tetralogy of Fallot and knowing the survival data for the total group, we derived survival curves for those presumed to have pulmonary stenosis (PS) rather than atresia (PA) as follows:

$$S_{CA}(t) = S_{PA}(t) + (1 - \alpha)S_{PS}(t) \quad (1)$$

where $S_{CA}(t)$ = survival curve for 566 composite autopsy (CA) cases; $S_{PA}(t)$ = survival curve for 54 cases of tetralogy of Fallot with pulmonary atresia; $S_{PS}(t)$ = generated survival curve for tetralogy of Fallot with pulmonary stenosis rather than pulmonary atresia; and $\alpha = 0.33$, the assumed incidence rate of pulmonary atresia at birth among patients with tetralogy of Fallot. Thus, the survival curve for the composite group is assumed to consist of the sum of the two separate survival curves proportionate to their respective incidence rates at birth. $S_{PS}(t)$ is derived by rearranging equation 1 algebraically.

Statistical methods: Nonparametric descriptions of the autopsy series were made using the product-limit method of Kaplan and Meier.[18] We modified their method to take into account the data from individual case reports as well as grouped data from which individual ages at death could not be determined (Hazelrig JB, Turner ME Jr, Blackstone EH, in preparation). Parametric estimates of survival (applicable to both the autopsy data and the Danish data) were calculated using the family of "survival equations" described by Turner and Pruitt.[19] The data were fitted to the "survival" equation using the method of maximal likelihood suggested by Gross and Clark[20] and a nonlinear optimization program developed by Hazelrig et al.[21] The specific survival equation selected from the family of equations was the one that did not fit the data significantly worse than the "generic" equation.

The parametric analysis yielded formulas that could be directly manipulated to generate the hazard function $\lambda(t)$, which is the instantaneous death rate:

$$\lambda(t) = -S'(t)/S(t) \quad (2)$$

where $S'(t)$ is the slope (first derivative) of the survival function $S(t)$. The formulas were also manipulated to generate the survival function for tetralogy of Fallot with pulmonary stenosis rather than pulmonary atresia, as described. The confidence limits for this calculated curve were computed from the variance (Var) estimated by the equation:

$$\mathrm{Var}[S_{CA}(t)] = \alpha^2\mathrm{Var}[S_{PA}(t)] + (1 - \alpha)^2\mathrm{Var}[S_{PS}(t)] \quad (3)$$

because $S_{PA}(t)$ and $S_{PS}(t)$ are independent. By algebraic manipulation, $\mathrm{Var}[S_{PS}(t)]$ could be computed.* The hazard function can be converted to a percent risk per year for a constant hazard rate by the formula $100 \times (1 - \exp[-\lambda])$, where exp is the base of the natural logarithms.

Results

The Danish study: According to the parametric analysis of the Danish study, 64 percent of patients born with tetralogy of Fallot, including those with pulmonary atresia, are alive at age 1 year, 54 percent at 2 years, 47 percent at 3 years and 24 percent at age 10 years (Fig. 1). The hazard function (or the instantaneous risk of death at any given age) is highest in the first year of life, gradually declines until age 10 years and remains essentially constant at about a 6.4 percent risk per year thereafter. Related to this is continual decrease in the number of survivors, so that only 11 percent of persons born with tetralogy of Fallot are alive at age 20 years, 6 percent at 30 years and 3 percent at 40 years.

Composite autopsy series: Parametric analysis of the composite autopsy data (566 cases) indicates that 66 percent of patients born with tetralogy of Fallot, including those with pulmonary atresia, are alive at age 1 year, 56 percent at 2 years, 48 percent at 3 years and 22 percent at age 10 years (Fig. 2). The hazard function is similar to that derived from the Danish study.

Combined series: Combined parametric analysis of the data from the Danish study and from autopsy reports (782 cases) is permissible because the survival function for the two series was not significantly different ($P = 0.2$). The combined analysis indicates that 66 percent of patients born with tetralogy of Fallot are alive at age 1 year, 56 percent at 2 years, 49 percent at 3 years and 24 percent at age 10 years (Fig. 3, A and B). The combined hazard function remains constant after age 10 years (Fig. 3C).

Tetralogy of Fallot with pulmonary atresia: The life expectancy of patients with tetralogy of Fallot and pulmonary atresia (54 cases) is shorter, according to the autopsy data (Fig. 4, A and B). By parametric analysis only 66 percent of patients are alive at age 6 months, 50 percent at 1 year, 33 percent at 2 years, 25 percent at 3 years and 8 percent at age 10 years. The 70 percent confidence limits are wider than in the previous analyses because of the smaller number of cases. The hazard function is very high in the first few years of life (Fig. 4C).

Tetralogy of Fallot without pulmonary atresia: The data from the autopsy study for all persons with tetralogy of Fallot known to be without pulmonary atresia (195 cases) are shown in Figure 5. The derived data (see Material and Methods) for persons with tetralogy of Fallot without pulmonary atresia are shown in Figure 6. The differences between the curves from the 195 cases and those from the derived data are largely in the first few years of life. The derived parametric analyses indicate that 75 percent of patients are alive at age 1 year, 60 percent at 3 years and 30 percent at age 10 years.

For completeness and ease of reference, the curves for persons with tetralogy of Fallot and those known to have pulmonary atresia and the *derived* curves for those without pulmonary atresia are shown in Figure 7. In general, persons with pulmonary atresia have the shortest life, and those with pulmonary stenosis have a better prognosis. The confidence limits of all survival curves overlap in the older age group, suggesting that there may be no significant differences among older patients in the three groups. In addition, the confidence limits of all curves overlap in the first 3 months of life.

Discussion

Evaluation of methods and their possible limitations: The Danish study assumed that 0.4 instances of the tetralogy of Fallot occurred in each 1,000 live births, and other studies[22–25] indicate that this assumption is reasonable. The similarity of the results in

* We are indebted to Dr. Edwin Bradley of the Department of Biostatistics for the development of these relations.

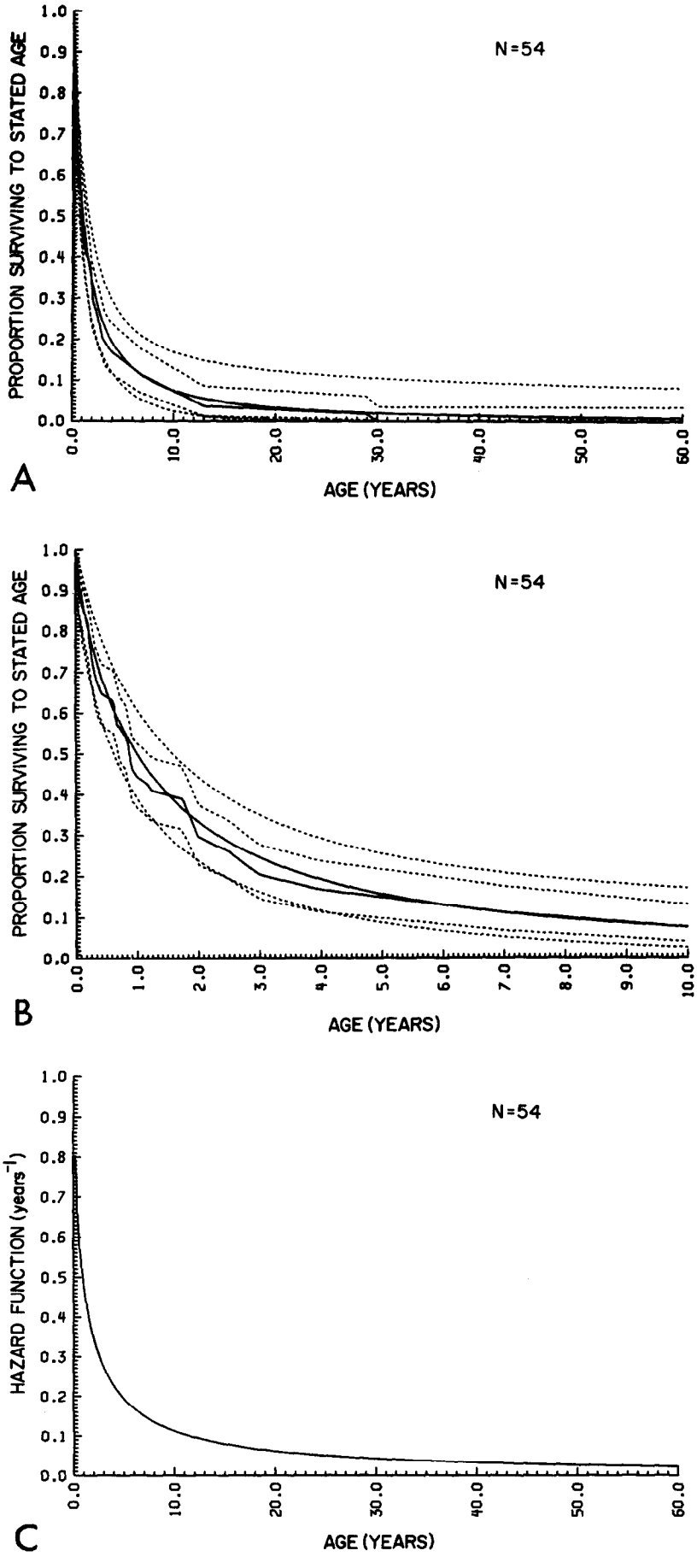


FIGURE 4. Life expectancy of persons with tetralogy of Fallot and known pulmonary atresia, based on autopsy data. The portrayal in **A, B** and **C** is as in Figure 3. The smooth line represents the equation (positive generic model[19]): Probability of dying = $[1 - (1 + \mu t/m\nu)^{-\nu}]^m$, where (as in Fig. 2) $\beta = -3.3 \pm 0.85$, t is age in months, $m = 0.7 \pm 0.22$ and $\nu = 1.4 \pm 0.80$. Significance level of coefficients: $P_\beta = 0.0003$, $P_m = 0.0001$ and $P_\nu = 0.09$ (chi square test for single hit model being different from this generic model, $P = 0.006$); correlation between coefficients, $r_{\beta,m} = -0.938$, $r_{\beta,\nu} = -0.697$ and $r_{m,\nu} = 0.558$. N = number of patients.

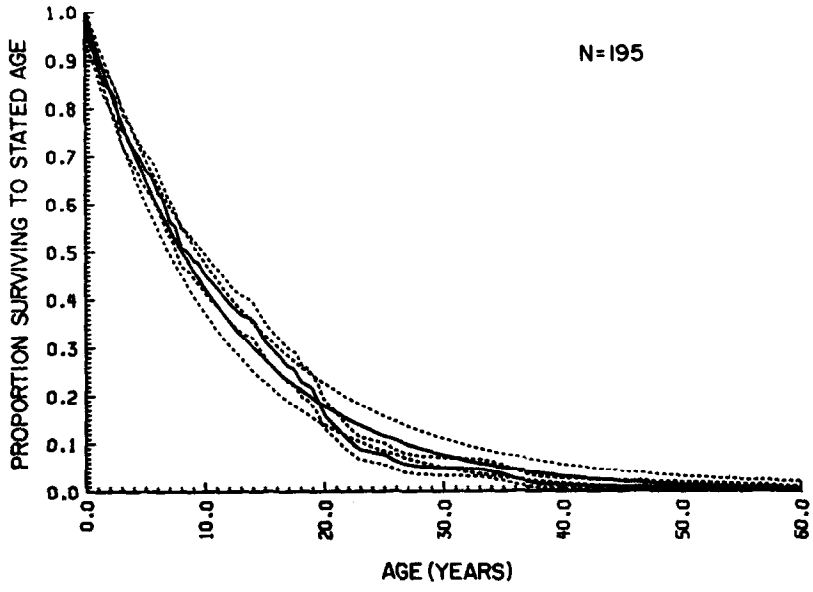


FIGURE 5. Life expectancy of persons with tetralogy of Fallot and known pulmonary **stenosis** (not atresia), based on autopsy data from 195 patients. The smooth line represents the equation: Probability of dying = $1 - \exp(-\mu t)$, where exp is e, the base of the natural logarithms; $\mu = \exp(\beta)$, where $\beta = -4.94 \pm 0.082$, and t is age in months. Significance level of coefficient, $P_\beta < 0.0001$. The **jagged line** represents the nonparametric actuarial analysis. N = number of patients.

the Danish study and in the composite autopsy study indicates that the cases in the latter are probably an adequate sample of persons born with tetralogy of Fallot.

The assumption that 33 percent of persons with tetralogy of Fallot have congenital pulmonary atresia is based on four reports indicating that this proportion is 27, 42, 39 and 35 percent, respectively.[14,22,26,27] This estimate is consistent with the recent experience of Arciniegas, who found 26 patients (22 percent) with tetralogy of Fallot and pulmonary atresia among the 118 patients with tetralogy on whom he operated in infancy (personal communication).

The nonparametric actuarial method used in the analyses is standard for handling this type of data, except for the important modification for grouped information. The method has several limitations. No interpolation scheme between data points at individual events or projection of confidence limits is theoretically admissible even when the actual data seem to follow a simple, smoothly decreasing pattern; in addition, confidence limits should widen as patients are withdrawn because of unknown status. A major weakness exists in handling a large number of patients "lost to follow-up" or "withdrawn alive," as in the Danish cross-sectional study for which no "actuarial" estimate is possible, in part because no events (deaths) were recorded. The theoretical binomial limits to any actuarial method in such cases are usually extraordinarily broad and may render any nonparametric method of little

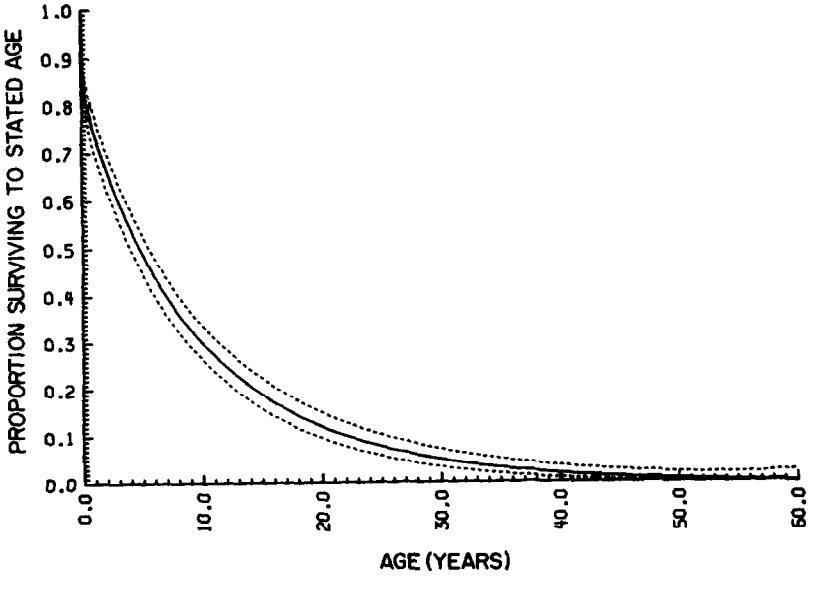


FIGURE 6. Life expectancy of persons with tetralogy of Fallot without pulmonary atresia, derived as described in Methods.

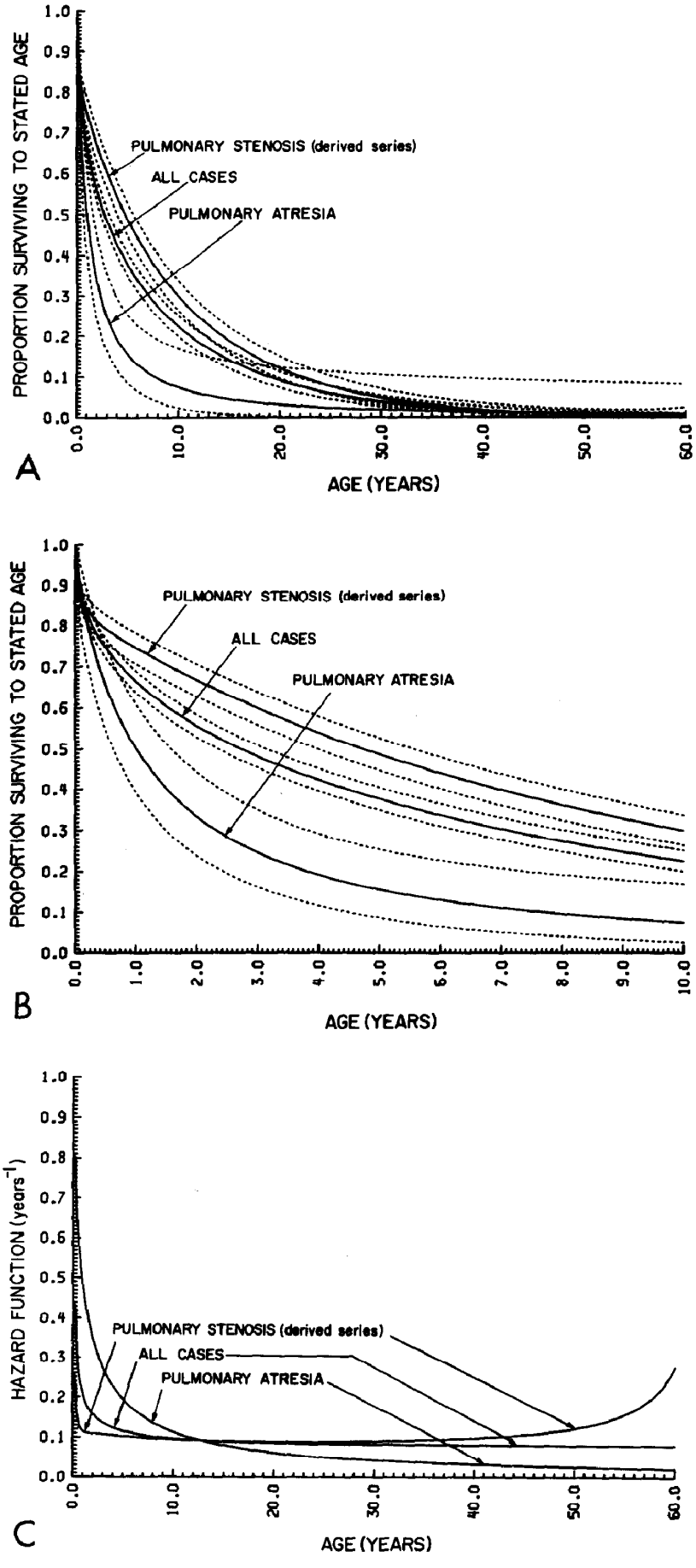


FIGURE 7. Combination of parametric equations from Figures 3, 4 and 6. The presentations in **A, B** and **C** are as in Figure 3.

value. Serial correlation of the actuarial estimates may produce errors throughout the calculated life table; the grouped data were especially troublesome in this regard, generating estimates that were at times outside the theoretical binomial limits.

The parametric technique overcomes these disadvantages by generating continuous estimates at all ages (as expected of any regression equation of low order); by treating "unknown" data as if they followed the expected survivorship curve for the known data, thereby allowing us to combine longitudinal information (autopsy series) with cross-sectional information (Danish series), which, as far as we know, has not previously been possible; and by treating individual events independently. In addition, the advantages of expressing a survivorship function as a simple formula have been recognized for many years, particularly in reports on industrial reliability.[20,28] With this method, the hazard function is easily calculated and provides insight into the changing risk among survivors. In addition, regression coefficients of different series can be compared so that statistical differences are easily determined, and the formulas can be manipulated to generate such useful functions as "surgical salvage," given the data on natural history and surgical risk. The *scheme* used in parametric analysis is unique in that a particular formula for the survivorship function is not assumed at the outset, but instead a flexible, high-order "generic" model is first fitted and then simplified, if possible, to one of the many formulas in the family of equations derived from the generic model.

Definition and material: We intended to include all persons with a morphologic diagnosis of tetralogy of Fallot—that is, all those with atrioventricular (A-V) concordant connection and a large ventricular septal defect in the left ventricular outflow tract immediately beneath the aortic valve, biventricular origin of the aorta, origin of the pulmonary artery from right ventricle and pulmonary stenosis that was at least partly infundibular and severe enough to produce marked right ventricular hypertrophy. Pulmonary atresia, when present, was pulmonary truncal, valvular, infundibular or valvular and infundibular, and the remnant of the pulmonary artery was above the right ventricle. The ventricular septal defect was subpulmonary in some cases. No case with complete A-V canal was included. We attempted to exclude cases with absent left and right pulmonary arteries, so-called truncus arteriosus type IV.[29]

We presume that the survival curves we derived for persons who have pulmonary atresia are applicable prognostically to persons who have atresia in the first few months of life, and that those derived for persons with pulmonary stenosis rather than atresia are applicable to persons who have tetralogy of Fallot with pulmonary stenosis in the first few months of life, whether or not they are cyanotic at that time or manifest pulmonary atresia later in life. The clinician must refine the prognostic prediction for an individual patient from these curves on the basis of severity of the patient's cyanosis and thus of the pulmonary stenosis.

We included in the autopsy group only patients believed to have died of causes related to their malformation. If the patient was known to have died of an unrelated cause such as tuberculosis or typhoid fever, the cause was excluded so that the survival data would be applicable at the present time.

Evaluation of role of severity of pulmonary stenosis in survival: The life expectancy of the group of patients with tetralogy of Fallot from the autopsy study is strikingly similar to that derived from the Danish study, in which a completely different study technique was used. This finding strongly suggests that both studies correctly predict the life expectancy of persons with tetralogy.

The data support the generally accepted concept that the natural history of persons born with tetralogy of Fallot is determined primarily by the severity of the pulmonary stenosis, as demonstrated by the tendency of persons with pulmonary atresia to die at a younger age than those without pulmonary atresia or the group as a whole. The infants with pulmonary atresia who die in the early months of life probably lack large discrete "bronchial" collateral arteries and die as a result of gradual spontaneous closure of the patent ductus arteriosus, which Bharati et al.[30] found in 81 percent of cases. Large "bronchial arteries" or, less commonly, large discrete paramediastinal vessels or coronary-pulmonary artery fistulas,[31] are present in many of the patients with tetralogy of Fallot and congenital pulmonary atresia (38 of 80 surgical cases in the experience of Alfieri et al.[32]) and do not close spontaneously. Although these vessels often allow the infant to survive the early months of life, the collateral pulmonary flow they provide is frequently insufficient to prevent arterial desaturation and cyanosis. Thus, many such patients die in childhood. Those who reach the third decade of life have a large collateral pulmonary blood flow and probably as a result are at that age at less risk of dying at any given time than persons with classic tetralogy of Fallot without pulmonary atresia.

In the subset of persons with tetralogy of Fallot and pulmonary stenosis, the pulmonary stenosis usually becomes more severe with time and results in increasing arterial desaturation, cyanosis, polycythemia and, ultimately death, usually from hypoxia, pulmonary thromboses or cerebral thromboses or abscesses. In patients surviving into the fourth and fifth decades of life, congestive heart failure may cause death.

Therapeutic implications: Because about one half of surgically untreated patients with tetralogy of Fallot die in the first 2 years of life, surgical programs for persons with this malformation must include techniques for surgical intervention in the early months and years of life. Also, because the hazard function does not decrease in the third to fifth decades of life, the person with tetralogy in this age group is still subject to the risks of the malformation.

Institutions reporting that less than half of their patients treated surgically for tetralogy of Fallot are younger than 2 years of age may be delaying surgical intervention unwisely or may have a patient population that represents less than the full spectrum of cases of tetralogy of Fallot.

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